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A FORM OF DISEASE HITHERTO UNDESCRIBED.

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*Reprinted from the
Transactions of the College of Physicians of Philadelphia, 1892,
and from
The American Journal of the Medical Sciences, May, 1892.*

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[Read February 3, 1892.]

IN April, 1891, I read before the College a paper entitled, "The Microscopic Anatomy of the Human Heart," which appeared in the TRANSACTIONS of the College of Physicians of Philadelphia, and was published besides in *The American Journal of the Medical Sciences* for June, 1891. It was then shown that in the human heart in the natural condition the muscular fibres are penetrated by capillaries, and that they are not therefore, as is commonly supposed, mere solid rods. At the same time, I alluded to the fact that I was able to make this observation owing to a certain pathological change that I had noticed in studying diseased hearts—a change by which the nearly solid normal fibres became tubes. So far as my observations extend, this condition is usually most marked in the fibres of the papillary muscles of the left ventricle, though it is common in all other parts of the heart as well. The degree of the excavation varies exceedingly; the cavities may be so small that in some instances it is impossible to distinguish them from capillaries, or, on the other hand, the hollowing-out process may have gone so far that the fibres are changed into tubes with thin walls. The disease may be best studied when the fibres are seen in cross-section, for then its most character-

istic appearances are presented. Its presence can, however, be equally positively determined in longitudinal sections, though greater care and discrimination must then be exercised to recognize it. The drawing (Fig. 1) represents types of the morbid changes as they appear when present in a high degree of development. There are two fibres which are natural; the others have been more or less eaten away in their centres by disease. The destructive process, in its most extreme form of development, removes the whole of the muscular substance from the centre of the fibre, no part of which, when examined with the microscope, will present the usual appearance of muscular tissue except the thin outer walls, and even these (Fig. 1, No. 7) may show only in places the cross-markings characteristic of heart muscle. A curious feature is the way in which the muscle nuclei often lie loosely in the cavities, without attachment to the remaining tissue. This is very different from the natural condition when the nuclei are closely surrounded by the muscular tissue. This separation of the nucleus was very marked in the two fibres represented in Fig. 1 as Nos. 5 and 7. Nos. 3 and 5, and, in a slightly different stage, No. 4, represent another phase of the destructive process. In them, instead of a single large cavity in the centre, there are several smaller holes irregularly distributed through the fibre, and in No. 5 the muscle nucleus lies in the largest of these. The degenerative process is more advanced in No. 4 than in the two others, for in it the only muscular tissue remaining is that constituting the thin outer walls, with a little more which is irregularly distributed through the inner portions, and some shreds of a material which must be the endomysium or fine connective-tissue substance which normally exists in the muscular fibres. It must be understood that these cavities do not usually give the impression that during life they were empty, or even that they contained only a clear liquid, for there is always present more or less material that has no distinguishable structure. Areas presenting this condition are represented in the drawings by the dotted portions. It seems to me that this hollowing-out, when most extreme, is apt to be near the ends of the

fibres, toward their point of origin or of insertion. In sections of tissue that show it I have found the hollow fibres most numerous not far from the endocardium or pericardium, and less so deeply in the centre, though, as already stated, it will be unmistakably present in all parts of the heart. The heart which furnished the fibres represented in Fig. 1 was very fibroid, but the hollow fibres were not found in the fibroid portions of it. Fig. 2 was made by the photographic process alone, without retouching, and shows fibres in varying stages of degeneration.¹ In it may be seen several of the same fibres as are shown in Fig. 1. In this heart, however, I was able to study the disease to the best advantage because it was present in its most extreme form.

The patient was a man fifty-seven years of age, who had been very dissipated, having eaten and drunk too much, and having done most of the things a man should not do. He denied ever having had syphilis, but at sixteen years of age he had an attack of inflammatory rheumatism. He said he had been healthy until recently (or about six months before his death), when he became exceedingly short of breath, and had some irregular rheumatism. His condition became gradually worse until he died, after having suffered to an unusually great degree with difficulty of breathing. For many weeks there was general oedema and ascites, also albumin and casts in the urine, and great atheromatous stiffening of the arteries. The post-mortem examination showed fibrosis of most of the organs, including the heart, which was much enlarged. The kidneys were contracted, and there was an extraordinary amount of chalky deposit, which was not confined to the walls of the arteries alone, for there were large deposits in the mesentery and in the posterior walls of the abdominal cavity.

The first case in which I discovered the presence of the morbid change in the heart-fibres was that of a man past middle life, who had died with aortic regurgitation. He denied having had rheumatism and syphilis, and died of exhaustion after an illness of a few months, without ever having had dropsy, or dyspnoea, or any evidence of kidney disease. There was aortic disease, and the heart was enlarged. The kidneys were rather larger than normal, and but slightly diseased. Excessive hollowing out of the muscular fibres of the heart was found.

¹ The photograph, which is a very beautiful one, was taken by Dr. George A. Piersol, Professor of Anatomy in the University of Pennsylvania.

It is impossible at present to predict from clinical manifestations which cases will present this curious change in the heart. Some idea, however, of its clinical and pathological accompaniments may be gained by contrasting cases in which the disease was present, with others not so diseased. As was stated in my paper showing the existence of capillaries in the heart's muscle-fibres, I have in my collection sections of forty-nine human hearts, accompanied in most instances by clinical histories of the patients. In some typical cases of Bright's disease, both of the interstitial (or contracting) and of the parenchymatous forms, hollow fibres were present, and in other like cases I have not found them. In hearts showing great increase of fibrous material, it has in some instances been present and in others absent. It is a curious feature that the vacuolations, when present in a fibroid heart, occupy almost exclusively those portions of the organ not affected by the fibroid overgrowth; in such regions it will be conspicuous by its absence. There are in my collection sections from two hearts showing more marked fibroid overgrowth than any of the others; in one, the hollowing-out of the muscular fibres is very great, in the other there is none of it; in both cases the kidneys were very much contracted. The history of one of these cases has been given in some detail above. Examination of such sections as are in my possession shows the vacuolation of the fibres to be absent in cases of brain syphilis, sarcoma, general miliary tuberculosis, Bright's disease, pulmonary phthisis, typhoid fever, pneumonia, dysentery, epithelioma of the bowel, and aneurism, and to be present in what clinically was recognized as organic heart disease, Bright's disease, typhoid fever, ulcerative endocarditis, and in young infants that had died of wasting. I have sections from the hearts of a number of foundlings who died during the earlier months of life from the wasting so common among infants of that class, and in some instances vacuolation of the heart's muscle-fibres was unmistakably present, and in others absent. In many respects the histological condition of heart muscle in young infants, as might be expected, is very different from what it is later in life. In a child ten years of age who died with

dropsy, and had amyloid disease of the liver, spleen, and kidneys, and of the heart and lungs too, there was most extreme hollowing-out of the muscle-fibres. Some of the fibres in this case, when seen in longitudinal section, presented small bulbous-looking swellings at points where they were hollow, and thus making it appear that some distending process had occurred where the vacuolations existed. In another child of twelve, with cardiac hypertrophy and dropsy, the vacuolations were also present, but in much less degree than in the previously mentioned case.

It has been my endeavor to describe this pathological change so that it may be recognized by others; and, as previously stated, it was while studying it that I was led to the discovery that capillaries normally penetrate to the very centres of the muscular fibres of the heart. The kind of cases in which the disease was found I have also denoted. It is now desirable to understand, if possible, the nature and origin of the morbid process. The fact that the muscular fibres of the heart are penetrated by capillaries, and are not, therefore, truly solid bodies, together with the appearance of the spaces already described, lead to the almost inevitable conclusion that the process is one of cystic degeneration. The only other conceivable explanation is that the cavities are minute aneurisms, being dilatations of the capillaries after they have passed into the muscular fibres. Such an assumption, however, would seem to be negatived by the nature of the material which lies within the cavities. Any material visible is amorphous and granular, or is yellowish pigment in irregularly shaped flakes, all looking as if suspended in a liquid, and thus presenting the precise characteristics of any section of a cyst. If the process be one of multiple capillary aneurism, fresh blood should be found in the spaces instead of the detritus described. Such being the case, it would seem hard to escape from the conclusion that the cavities are true retention cysts, and that they are produced in a manner parallel to that of renal cysts. A capillary must become blocked in two places and the portion between these dilate, and thus a cyst be formed. It is, of course, quite possible that the

vaeuulations are false eysts formed by the eseape of blood into the substance of the fibres (haematocele), or by degeneration and softening of the muscle-substance itself. This, though quite admissible as a possible explanation of the pathological process, is much less likely to be the real one than is the supposition that they are true retention cysts ; for, in the first place, there are in the fibres cavities, the capillaries, in which cysts might form, and, in the second, the contents are precisely similar to those common to eysts. These cavities present another characteristic which is common also in renal cysts. Shreds, or festoons, or shelf-like projections of connective tissue hang irregularly across the cavities. Nos. 3 and 4, Fig. 1, exhibit this appearance.

If time and the progress of events should establish that these minute cavities in the heart's muscle-fibres are true retention cysts, having their origin within capillaries in the fibres, it will be the first time that it has been shown that eysts do originate within the vascular channels, as they are already so well known to originate within other channels, as, for instance, the renal tubules and other gland ducts. There seems to be no reason in the nature of things why such should not be the case.

The presence of capillaries within the muscular fibres of the heart, a fact in normal anatomy, and the occurrenee of cysts within the fibres, a fact in pathologieal anatomy, and the fact that the one observation led to the other, are an illustration of the correctness of the view so much dwelt upon now, that the two branches of science, normal and pathological histology, have close and important relations. In this instance the train of events was as follows: First, as the result of pathological study, the recognition of the comparatively large cavities in the fibres, which at that stage were inexplicable. From this came the desire to examine more minutely the histology of the human heart, and this resulted in the observation that the capillaries not only closely surround the fibres, but that they actually penetrate them, a fact which seemed at first in no way connected with the holes eaten into the fibres by disease. Lastly, further

and more careful consideration leads to the conclusion that the larger spaces which result from disease are but cystic dilatations of the natural blood channels.

It is important to remember that this disease, which I have called cystic degeneration, is very common, and has been found by me to be present in many ordinary well-recognized diseases. What bearing it may be found to have clinically is, of course, impossible now to predict, but it seems not unlikely that it may be important. The thoughts certainly are new ones, both that cystic disease may have its origin in the vascular system, and that it is a common cause of degeneration of the heart, an organ which is so prone to degenerate, and degeneration of which is so far-reaching in its results.

NOTE.—It should have been mentioned that the sections of tissue from the appearances of which my conclusions were drawn were prepared as far as possible, upon a uniform plan, both for the preservation of the tissue and the mounting of the sections. I thought that if this was done the unnatural appearances could with more certainty be attributed to disease than if different methods were used, in which case it might be thought that what I described as disease was due to faulty technique. The tissues were almost all preserved in seventy per cent. alcohol, and the imbedding material was paraffine, except in a few instances, when eelloidin was used. The staining material was borax carmine. Considering the imbedding material employed, and that the sections were often cut from parts of tissue far from the surface of the pieces, it would be unreasonable to suppose that what I have called hollow muscle-fibres were fibres from which the centres had fallen out in process of preparation, a possibility which would strike every practical microscopist.

EXPLANATION OF FIGURES.

FIG. 1.—The Nos. from 1 to 6 inclusive represent heart muscle-fibres in cross-section, and were all drawn from the same microscopical preparation. No. 7 was drawn from a preparation in which the fibres were cut longitudinally. The camera was used, and they were drawn to scale, the amplification being five hundred diameters. Nos. 1 and 2 are ordinary fibres, and present no evidence of disease; they were put in the picture to contrast with the other fibres, which are diseased. The irregular shape of the nucleus shown in No. 2 is not unusual. The other fibres present evidences of advanced disease. Nos. 3, 4 and 6 look as if irregularly hollowed out, the dark areas being intended to represent the muscular tissue and endomysium (intramuscular connective tissue), while the lighter portions are areas from which the muscle-substance has disappeared as a result of disease. No. 5 is a large fibre, showing several spaces from which the muscle-tissue has gone, the space being empty, or filled with a more or less granular material. There is also a nucleus which lies in a space which is empty or filled with slightly granular matter, this being represented in the drawing by a dotted area. No. 7 is a drawing of a fibre cut longitudinally, and very well shows the irregular hollowing-out process or vacuolation which has taken place. The fibre looks as if it had been converted into a hollow cylinder divided into several compartments. The drawing was made to show that at the sides of the fibre the muscular tissue still persists, in places the cross striæ even being distinguishable, while the dots in the central portion are intended to indicate the more or less granular appearance. The nuclei lie as if they had been loose in the central space, not being in any part in contact with the muscular tissue remaining. The drawings are diagrammatic, but the dimensions and outlines are correct.

FIG. 2.—This is a photograph (a half-tone) of a single field under the microscope of the papillary muscle of the left ventricle from the same case as that from which the drawings were made. Several of the fibres in the drawing are in this picture. It admirably shows the manner in which the interior of the fibres has been irregularly eaten away by disease. Neither the negative nor the print from which this was made was retouched.

FIG. 1.

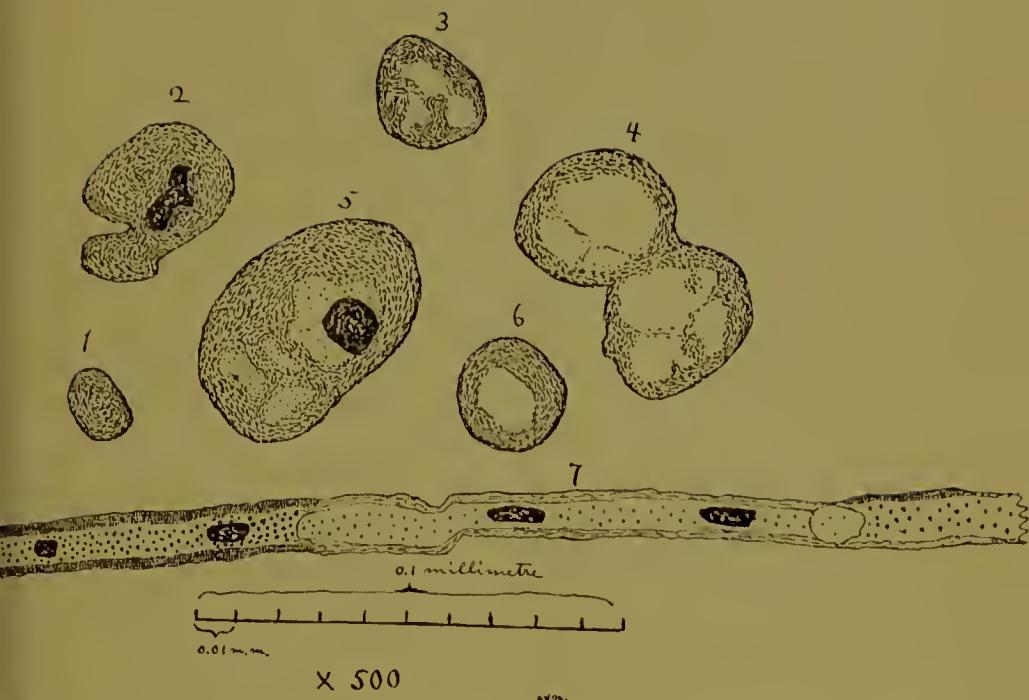


FIG. 2.



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